NEWS RELEASE

Early and Sustained Increase in Serum TTR Levels by Acoramidis Independently Predicted Improved Survival in the ATTRibute-CM Study

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- For each 5-mg/dL increase in serum TTR level within 28 days of starting treatment, the relative risk reduction of mortality was up to 31.6% through Month 30, confirming the hypothesis that ever better levels of stabilization achieved by treatment with acoramidis, a near-complete (≥90%) TTR stabilizer, lead to ever better clinical outcomes

- ATTRibute-CM is the only study to demonstrate a direct association between a prompt, sustained increase in serum TTR and survival in patients with ATTR-CM

- The open-label extension data for all ATTRibute-CM participants at Month 42 showed that rapid, sustained TTR stabilization from acoramidis demonstrated statistically significant reductions in ACM and CVH (including urgent outpatient treatment for heart failure exacerbations)

- In the ATTRibute-CM study, acoramidis demonstrated the most rapid benefit seen in any Phase 3 study of ATTR-CM to date in both ATTRv-CM and ATTRwt-CM patients:

    - In as few as 3 months, the time to first event (ACM or CVH) durably separated relative to placebo
    - A 42% reduction in composite ACM and recurrent CVH events relative to placebo at Month 30
    - A 50% reduction in the cumulative frequency of CVH events relative to placebo at Month 30

- Acoramidis is approved as Attruby™ by the U.S. FDA and is approved as BEYONTTRA® by the European Commission, the Japanese Pharmaceuticals and Medical Devices Agency, and the UK Medicines and Healthcare

Products Regulatory Agency

PALO ALTO, Calif., May 19, 2025 (GLOBE NEWSWIRE) -- BridgeBio Pharma, Inc. (Nasdaq: BBIO) (“BridgeBio” or the “Company”), a new type of biopharmaceutical company focused on genetic diseases, published data showing that an early, sustained increase in serum transthyretin (TTR) levels predicted improved survival in ATTRibute-CM, its Phase 3 trial of acoramidis in transthyretin amyloid cardiomyopathy (ATTR-CM). These findings were published in Journal of the American College of Cardiology (JACC) in the Special Focus Issue: Amyloid. Acoramidis is a selective, small molecule, orally administered, near-complete (≥90%) TTR stabilizer. These findings further support the thesis that ever better increases in serum TTR lead to ever better clinical outcomes and that early elevations in serum TTR are an important prognostic marker to inform treatment selection.

“Patients with ATTR-CM have progressive amyloid accumulation in the heart, which when untreated manifests as progressive heart failure, arrhythmias, and eventually can result in death. Increases in serum TTR seen with acoramidis therapy within 28 days of initiation and that were sustained with therapy, were associated with a decrease in all-cause mortality independent of baseline risk among subjects in the ATTRibute-CM trial. The increase in serum TTR is hypothesized to be due to a leftward shift in amyloidogenic TTR to a more stable tetrameric TTR. This is the first concrete evidence that there is a link between this rapid increase in serum TTR and survival. Such data may inform clinical practice, as early and sustained increases in serum TTR could represent a new potential ATTR disease-specific and prognostic biomarker that may further inform clinical decisions in optimizing care for ATTR-CM patients,” said Mathew Maurer, M.D. of Columbia University Irving Medical Center.

In patients with ATTR-CM, aging or an inherited variant can cause tetrameric TTR to destabilize and misfold, causing buildup of amyloid fibrils in the organs, specifically in the heart. These data demonstrate that by using acoramidis, a selective near-complete TTR stabilizer to bind serum TTR, a rapid and sustained increase in tetrameric TTR was independently associated with an improvement in overall survival, even after adjustment for known predictors like TTR variant status, baseline New York Heart Association (NYHA) functional class, baseline National Amyloidosis Centre (NAC) stage, and baseline serum TTR levels. Findings from the analysis included:

- Treatment with acoramidis resulted in a sharp, significant early rise in serum TTR levels (mean 9.1 mg/dL) within 28 days which was sustained throughout the 30-month treatment period
- For every 5-mg/dL increase in serum TTR level, the Cox proportional hazards model predicted a relative risk reduction of mortality of 26.6% and the logistic model predicted a relative reduction of 31.6% in odds of death through Month 30
- An early increase in serum TTR levels on Day 28 of dosing was associated with reduced all-cause mortality (ACM) in univariate analysis, an association which persisted in multivariate analysis independent of TTR variant status, baseline NYHA functional class, baseline NAC stage, and baseline serum TTR levels

- Logistic modeling demonstrated that among participants treated with acoramidis, the early increase in serum TTR was associated with reduced ACM, whereas there was no prompt and sustained increase in serum TTR observed in participants treated with placebo
- Causal mediation analysis showed evidence that the acoramidis treatment effect on ACM probability through month 30 was fully mediated by the observed prompt and sustained increase in serum TTR

“This landmark analysis adds an incredibly important proof point to acoramidis’s repository of compelling data that higher serum TTR levels are directly correlated with and mediate a reduction in mortality risk.” said Jonathan Fox, M.D., Ph.D., Chief Medical Officer of BridgeBio Cardiorenal. “We believe that this will be an important measure for physicians treating ATTR-CM to consider and will be encouraging information for new and existing patients when reviewing options to treat their condition.”

Data from 42 months sustained treatment in the open-label extension study from ATTRibute-CM showed that rapid and sustained TTR stabilization from acoramidis demonstrated statistically significant reductions in both ACM and cardiovascular-related hospitalizations (CVH, which included urgent outpatient treatment for heart failure exacerbations). The continued curve separation of the composite endpoint of ACM and CVH emphasizes the importance of early and continuous treatment resulting in early and sustained clinical benefits. These data further underscore the hypothesis that ever better levels of stabilization lead to ever better clinical outcomes and emphasizes the importance of a prognostic biomarker, serum TTR, to inform decision making for patient care.

Acoramidis is approved as Attruby by the U.S. FDA and is approved as BEYONTTRA by the European Commission, Japanese Pharmaceuticals and Medical Devices Agency, and UK Medicines and Healthcare Products Regulatory Agency with all labels specifying near-complete stabilization of TTR.

About Attruby™ (acoramidis)
INDICATION
Attruby is a transthyretin stabilizer indicated for the treatment of the cardiomyopathy of wild-type or variant transthyretin-mediated amyloidosis (ATTR-CM) in adults to reduce cardiovascular death and cardiovascular-related hospitalization.

IMPORTANT SAFETY INFORMATION
Adverse Reactions
Diarrhea (11.6% vs 7.6%) and upper abdominal pain (5.5% vs 1.4%) were reported in patients treated with Attruby versus placebo, respectively. The majority of these adverse reactions were mild and resolved without drug discontinuation. Discontinuation rates due to adverse events were similar between patients treated with Attruby versus placebo (9.3% and 8.5%, respectively).

About BridgeBio Pharma, Inc.

BridgeBio Pharma, Inc. (BridgeBio) is a new type of biopharmaceutical company founded to discover, create, test, and deliver transformative medicines to treat patients who suffer from genetic diseases. BridgeBio's pipeline of development programs ranges from early science to advanced clinical trials. BridgeBio was founded in 2015 and its team of experienced drug discoverers, developers and innovators are committed to applying advances in genetic medicine to help patients as quickly as possible. For more information visit **bridgebio.com** and follow us on **LinkedIn**, **Twitter, Facebook**, and **YouTube**.

BridgeBio Forward-Looking Statements

This press release contains forward-looking statements. Statements in this press release may include statements that are not historical facts and are considered forward-looking within the meaning of Section 27A of the Securities Act of 1933, as amended (the "Securities Act"), and Section 21E of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), which are usually identified by the use of words such as "anticipates," "believes," "continues," "could," "estimates," "expects," "hopes," "intends," "may," "plans," "projects," "potential," "seeks," "should," "will," and variations of such words or similar expressions. BridgeBio intends these forward-looking statements to be covered by the safe harbor provisions for forward-looking statements contained in Section 27A of the Securities Act and Section 21E of the Exchange Act. These forward-looking statements, including statements regarding the potential for sustained increases in serum TTR to serve as a disease-specific and prognostic biomarker in ATTR-CM, the belief that early elevations in serum TTR are an important prognostic marker to inform treatment selection, the hypothesis that improved serum TTR stabilization is correlated with better clinical outcomes, and expectations about how these findings may guide physician decision-making and patient care, reflect BridgeBio's current views about its plans, intentions, expectations, and strategies, which are based on the information currently available to BridgeBio and on assumptions it has made. Although BridgeBio believes that its plans, intentions, expectations, and strategies as reflected in or suggested by these forward-looking statements are reasonable, it can give no assurance that such plans, intentions, expectations, or strategies will be attained or achieved. Furthermore, actual results may differ materially from those described in the forward-looking statements and will be affected by a number of risks, uncertainties, and assumptions, including, but not limited to: the risks associated with BridgeBio's dependence on third parties for development; regulatory authorities requiring additional studies or data to support the continued or expanded commercialization of acoramidis; whether data and results meet regulatory requirements or are sufficient for continued development, review, or approval; and whether other regulatory agencies agree with BridgeBio's strategies or data interpretations. These risks also include impacts from global health emergencies, such as delays in regulatory reviews and other activities, manufacturing and supply chain interruptions, adverse effects on healthcare systems, and disruption of the global economy; and the impacts of macroeconomic and geopolitical events, including changing conditions from hostilities in Ukraine and in Israel and the Gaza Strip, increasing inflation rates, and fluctuating interest rates on BridgeBio's operations and expectations. Additional risks are described in the Risk Factors section of BridgeBio's most recent Annual Report on Form 10-K,

Quarterly Report on Form 10-Q, and other filings with the U.S. Securities and Exchange Commission. Moreover, BridgeBio operates in a very competitive and rapidly changing environment in which new risks emerge from time to time. These forward-looking statements are based upon the current expectations and beliefs of BridgeBio's management as of the date of this press release and are subject to certain risks and uncertainties that could cause actual results to differ materially from those described in these statements. Except as required by applicable law, BridgeBio assumes no obligation to publicly update any forward-looking statements, whether as a result of new information, future events, or otherwise.

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